

## POSTEROLATERAL DIAPHRAGMATIC HERNIA – THE HYDERABAD EXPERIENCE

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### ABSTRACT

**OBJECTIVES:** To see the mode of presentation and treatment of posterolateral diaphragmatic hernia and the complications and mortality associated with it in our setting.

**DESIGN:** A descriptive study conducted during October 1998 to April 2002 at Liaquat Medical University Hospital Hyderabad, Pakistan.

**METHODS:** A retrospective review of charts was done and 13 patients with posterolateral diaphragmatic hernias (bochdalek hernia) were identified.

**RESULTS:** There were 11 girls and 2 boys. The average age of presentation was 23 hours and the referral was late in the home-delivered babies as compared to the hospital-delivered neonates. The common referring diagnosis was pneumonia, respiratory distress and congenital heart lesions. Eight neonates had symptoms 12 hours after the birth and five reported after first 24 hours of their life. Ten children had associated anomalies, of which six had multiple and four of them died without any surgical intervention. Nine children were operated after preoperative stabilization via the abdominal approach. In 89%, a direct closure of defect was done and in five, ventral hernia was created to accommodate the herniated abdominal viscera. Sac was identified in four cases and excised before closure of defect. Chest drain was put in five patients. Six children had various complications, which include pneumothorax, chest and wound infections. The seven children survived and the overall mortality was 46%.

**CONCLUSION:** Although, the results in the present set of available facilities are comparable with other local series, but an early diagnosis by providing education about such conditions at primary level, better pre and post-operative support and the facilities of neonatal intensive care unit will significantly help in the reduction of morbidity and mortality among these children.

**KEY WORDS:** Posterolateral diaphragmatic hernia. Bochdalek hernia. Diaphragmatic abnormalities. Children. Pakistan.

### INTRODUCTION

In utero, diaphragm develops from four analogue: septum transversum, the paired pleuroperitoneal membranes, mesenchyme of the esophagus and from the body wall.<sup>1</sup> Bochdalek or posterolateral diaphragmatic hernia occurs when the pleuroperitoneal membrane fails to fuse with the developing muscles of the diaphragm, thus leaving a weak or unfused part, which allows the herniation of intra-abdominal viscera (stomach, small intestine, colon, spleen or liver) into the chest.<sup>2</sup> (Fig-1) This occupation affects the development of lung, resulting in pulmonary hypoplasia, increased pulmonary resistance and reactive pulmonary hypertension. The anomaly is usually associated with other congenital malformations<sup>3,4</sup> and these two factors have a direct relationship with the clinical course and outcome of

these babies.

Bochdalek hernia occurs in 1 in 5000 live births with great incidence in stillbirths and abortions.<sup>5,6</sup> It is also one of the acute pediatric emergencies and an important cause of perinatal mortality.<sup>4</sup> Defects are more common on left (80%), than right side, and sometimes occur bilaterally.<sup>7</sup> Usually the majority of these children present in first 24 hours of life, however, some may present in later age.<sup>8,9</sup> The babies, who present within first few hours after the birth often have more severe degree of pulmonary hypoplasia and reactive pulmonary hypertension, thus are 'high-risk' with the worst prognosis. The mortality rate in these high-risk patients remain still very high, even with modern inventions like extracorporeal membrane oxygenation (ECMO), high frequency oscillatory ventilation (HFOV), and inhaled nitric.<sup>10-12</sup> The babies who present late usually have less severe

defects and a better survival than the others, when managed properly in the time.

There is lack of contemporary data about the prevalence, clinical details, management and the outcome of this entity at the national level and to date only one series and few published case reports are available for review.<sup>9,13,14</sup> This study presents our experience with posterolateral (bochdalek) hernia.

## METHODS

This descriptive study was conducted during October 1998 to April 2002 at Liaquat Medical University Hospital Hyderabad. During this period, the medical

records of all patients diagnosed with posterolateral diaphragmatic hernia were retrospectively reviewed and the data related to age, sex, birth weight, associated anomalies, onset of symptoms, time of referral, methods of diagnosis, treatment and mortality was studied. All the ethical considerations were also taken into account.

## RESULTS

Females were predominant (81%) as compared to males (**Fig-2**). Nine (70%) children were born at home and four (30%) were delivered in the different maternity homes and hospitals. One mother had

**Table I: Clinical data of children with bochdalek hernia of diaphragm**

No.	Age	Sex	Presentation	Del	Side	Diagnostic means	Surgical treatment	AM	Outcome
1	1 ½ d	F	Respiratory distress	HM	L	Plain X-ray chest	Laparotomy, direct closure of defect	No	Survived
2	2 hrs	F	Cyanosis and respiratory distress	HO	L	Plain X-ray chest	No Surgical Intervention	Yes	Expired
3	18 hrs	F	Respiratory distress and dusky color	HM	L	Plain X-ray chest	Laparotomy, direct closure of defect	Yes	Survived
4	1 day	M	Respiratory distress	HM	L	Plain X-ray chest	Laparotomy, direct closure of defect	Yes	Survived
5	8 hrs	F	Cyanosis and respiratory distress	HO	L	Plain X-ray chest	Laparotomy, closure of defect by inner muscular flaps	Yes	Expired
6	4 hrs	F	Cyanosis and respiratory distress	HO	L	Plain X-ray chest	No Surgical Intervention	Yes	Expired
7	1 day	M	Respiratory distress	HM	L	Plain X-ray chest	Laparotomy, direct closure of defect	No	Survived
8	22 hrs	F	Respiratory distress	HM	L	Plain X-ray chest	Laparotomy, direct closure of defect	Yes	Survived
9	10 hrs	F	Cyanosis and respiratory distress	HO	L	Plain X-ray chest	No Surgical Intervention	Yes	Expired
10	12 hrs	F	Respiratory distress	HM	L	Plain X-ray chest	No Surgical Intervention	Yes	Expired
11	16 hrs	F	Cyanosis and respiratory distress	HM	L	Plain X-ray chest	Laparotomy, direct closure of defect	Yes	Expired
12	2 day	F	Respiratory distress	HM	L	Plain X-ray chest	Laparotomy, direct closure of defect	No	Survived
13	3 day	F	Respiratory distress	HM	L	Plain X-ray chest	Laparotomy, direct closure of defect	Yes	Survived
Del = Delivery HM = Home Delivery HO = Hospital Delivery AM = Associated Anomalies									

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routine antenatal ultrasound, but the defect was not picked up. The mean birth weight was 2800 gm (range 1200 – 3300 gm) and 23% (n=3) were younger than 36 weeks of gestational age. The average age at the time of presentation was 23 hours (range 2 – 72 hours) and the common presentation was respiratory distress and cyanosis. The clinical data is summarized in **Table I**. Ten children (77%) had associated anomalies and of these, 60% had multiple malformations (**Table II**). The diagnosis was made with the help of history of respiratory distress, cyanosis, scaphoid abdomen, chest auscultation and plain x-ray chest (**Fig-3**). Nine children were operated and an abdominal approach was made in all cases for the closure of defect. The sac was present in four cases and a direct closure of the hernia was done in 89% (n=8) cases. Ventral hernia was created in 56% (n=5) neonates. Chest drain was used in five patients for 24-28 hours. Six children had postoperative complications listed in **Table III**. The overall survival was 54% (n=7).

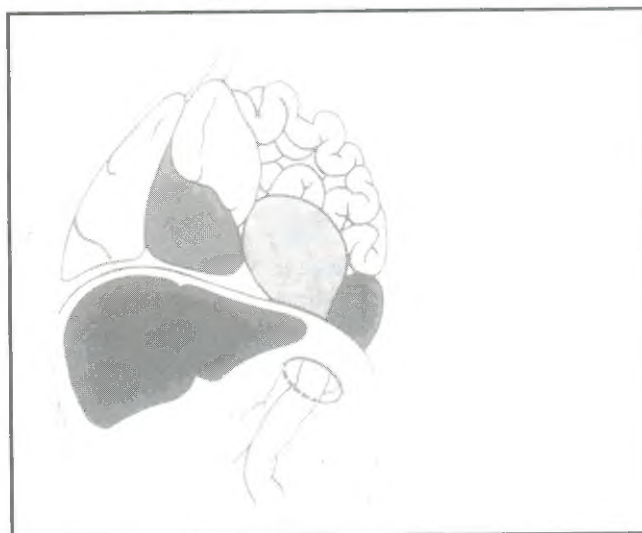
**Table II: Associated anomalies in children with congenital hernia of diaphragm (n=13)**

Anomalies	No.
VSD	5
ASD	3
Dextrocardia	3
Meningocele	1
Down Syndrome	2
Hypospadias	2
Cryptorchidism (Bil)	2
Amelia (lower extremity)	1
Malrotation of gut	1

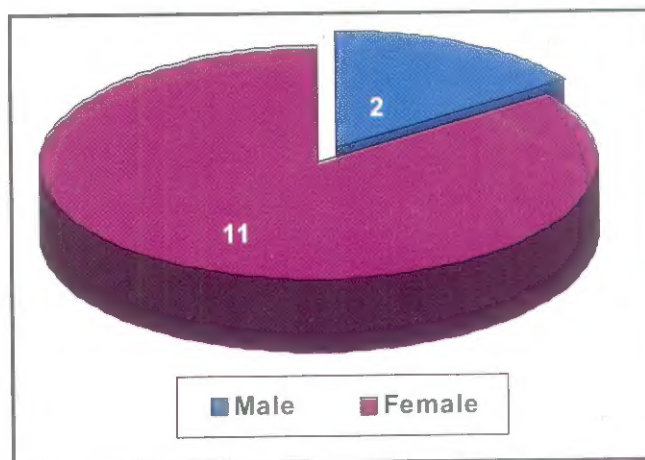
**Table III: Complications in children associated with congenital hernia of diaphragm (n=13)**

Complication	No.
Wound Infection	2
Chest Infection	2
Pneumothorax, ipsilateral	1
Pneumothorax, contralateral	1

**Figure 1: Schematic diagram showing bowel loops through posterolateral defect in the left hemithorax, with heart and left lung pushed to the right side**



**Figure 2: Sex Distribution of Cases**



**Figure 3: Plain x-ray chest showing bowel loops in left hemithorax with lack of demarcation of diaphragm**

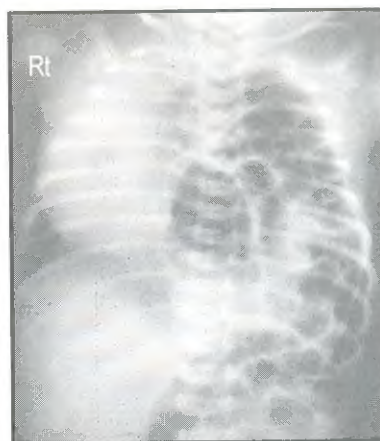


Figure 4: No. of Cases Per annum (1998-2002)

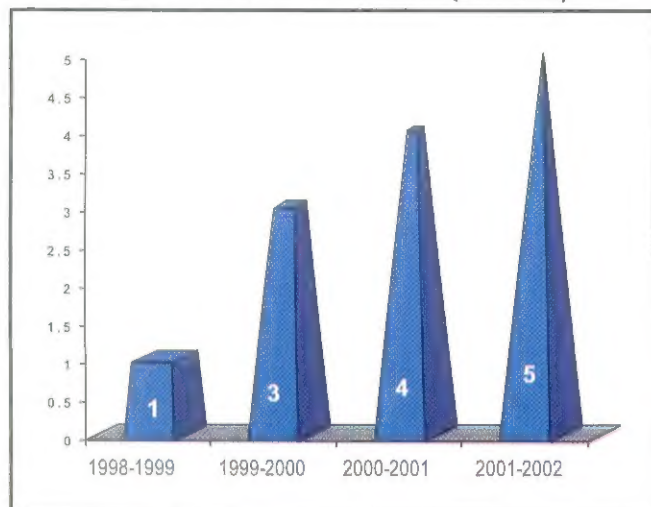


Figure 5: Operative view of posterolateral diaphragmatic defect



Figure 6: Post-operative view of the direct repaired posterolateral diaphragmatic defect



## DISCUSSION

The first description of congenital diaphragmatic hernia was reported as an incidental finding at postmortem examination and the first successful repair of this anomaly in a neonate of less than 24 hours of age was performed by Gross in 1940.<sup>15</sup> Since then, this anomaly is a common challenge for the team involved in the care of children as it is not only one of the acute pediatric emergencies but carries very high morbidity and mortality.<sup>12</sup>

The prevalence of bochdalek hernia varies from 1 in 2000 to 1 in 5000<sup>4,16</sup> but to assess this in our set up is bit hard because of lack of national register for the congenital malformations. In our institute, although, the reported cases with this entity increased with passage of time (**Fig-4**), but the overall occurrence was less than other reported congenital malformations. This may be because of lack of knowledge about the condition at primary level and also the non-availability of proper transportation for such babies from rural areas to the hospital. Thus, it is possible that a number of babies born with this entity may fail to get diagnosed and referred to the tertiary care units. The education about such paediatric surgical conditions at the primary level and better transportation facilities for these neonates will be helpful to pick more cases as well as to provide them proper treatment.

In our study, the lesion was common in girls with a male to female ratio of 1:4.5, which differs from the findings of Butler and David, who reported a ratio of 1:1.8 in their series.<sup>3,4</sup> Hernia is more common on the left side than right<sup>12</sup> and there was same observation in this study. The association of congenital anomalies with this entity is very common and the reported incidence of these malformations varies from 20% to 39% of live-born infants.<sup>4,17</sup> In our study, 77% (n=10) babies had associated congenital anomalies, of which six had multiple anomalies. The newborns with associated cardiac anomalies had more severe symptoms, early presentation and high mortality (**Table I**). As cited by Schumpelick et al, the diaphragmatic hernia, coupled with other major congenital anomalies, seems to always result in death.<sup>18</sup> and this was true in our study too, as four neonates, who died before any surgical intervention had other major anomalies, which contributed directly to the outcome.

The time of presentation after birth was variable ranging from 2 hours to 72 hours (average 23 hours). It was related with place of delivery, index of suspicion and onset of symptoms. Eight newborns developed symptoms 12 hours after the birth and 40% reported after first 24 hours of their life. 70% (n=9) neonates were born at home and of these five

were in the rural area. The diagnosis was established 'late' in home-delivered babies as compared to the hospital-born children. Although, the onset of symptoms was early in home-born babies, but the delay in the diagnosis was mainly due to the lack of knowledge about this condition among the health care practitioners. The main referral diagnosis for these children to our paediatric department was pneumonia, respiratory distress and congenital heart lesions. This finding put emphasis on the need of dispelling the education about paediatric surgery at various levels among the health care professionals especially in rural areas.

The diagnosis of the bochdalek hernia is generally made soon after birth on account of respiratory distress, cyanosis and a scaphoid abdomen. Usually a plain x-ray of the chest confirms this by showing heart and mediastinum shifted to the contralateral side and presence of bowel loops in the thoracic cavity with a lack of demarcation of diaphragm.<sup>19</sup> (Fig-3) In all our cases we reached the diagnosis by history, physical examination and plain radiograph of chest. Ultrasound and echocardiography were useful in the evaluation of associated congenital malformations of other systems. Once the patient was diagnosed, the stabilization was commenced by nasogastric tube for gastric decompression, vascular access, oxygenation and supportive medical treatment. With these manoeuvres, we achieved the optimal stabilisation before surgery in majority of cases, but the patients who required mechanical ventilation were referred as this facility was not available in our set up.

In the 1970s, this condition was an urgent indication for the surgical intervention as it was thought that the abdominal contents should be returned to the abdomen as soon as possible to allow the lungs to expand. Since early 1980, the situation has changed and today a child with this lesion is operated only when he/she is stable.<sup>20,21</sup> Clark et al in a multicenter study (62 centres with 461 patients), has reported that the average age of newborn at the time of operation was 73 hours (range 1-445 hours).<sup>12</sup> In our institute, we operated cases once they were stabilized with fluids, plasma, drugs and oxygen, and the average age for operation was 38 hours (range 6-98 hours). The recommended approach for the repair of such defects is transabdominal as it facilitates reduction of bowel, allows stretching of the abdominal wall to accommodate the bowel, increases the abdominal capacity by making a ventral hernia and facilitates additional procedure, if required.<sup>22</sup> In our series, we also operated all our cases via abdominal route taking the advantage of making ventral hernia in five (56%) patients because of their small abdominal cavity to accommodate the herniated viscera from

chest and also because of lack of the facility of mechanical ventilation. This worked well and we managed to save these babies with this maneuver. In one patient, who also had associated malrotation of gut, transabdominal approach was beneficial as we corrected this at the same time. Sac was present in 44% of our cases and it was excised before the closure of defect. The defects were variable in size in each child ranging from small/medium size defects (n=8) to large size defects (n=1). (Fig-5) We were successful in making a direct closure of medium size defects in 89% (n=8) cases (Fig-6), but in the child with large defect, the hernia was repaired with the help of pedicled abdominal wall muscle flaps as we lacked the facility of having synthetic mesh. We used chest drain in first five cases, but avoided in last 3 cases and no significant difference was noted between two groups in postoperative period. Similar findings are reported by Wung et al<sup>23</sup> where they managed their sixty-three cases without chest tube drainage and had good results.

Morbidity is related with the patient's pulmonary and cardiovascular status, and also with preoperative stabilization. Various authors have reported a complication rate of 44-63% in their series.<sup>24-26</sup> In our series six patients (66%) had various complications, which include pneumothorax (2), wound infection (2), and chest infection (2). None of these contributed in the mortality. The two children developed the pneumothorax after surgery, which required emergency chest tube insertion to save their lives. Persistent chest infections and wound sepsis were most likely secondary to post-anesthesia and poor general status, which responded well to the appropriate course of antibiotics after culture of organisms. The overall mortality rate in our study was 46% (n=6). Four neonates died before any surgical intervention and all these had associated cardiac malformations, which contributed directly to their death. Also, these patients had early onset of the symptoms (with in first few hours of their life), which is suggestive of severe pulmonary hypoplasia and reactive pulmonary hypertension. The other two babies also had multiple associated anomalies along with posterolateral defect, but these were not fatal and also the onset of symptoms in these two was late. The child with large defect who had repair with the muscular flaps had a difficult post-operative course and she died 18 hours after surgery. Another child died 10 hours after surgery because of respiratory insufficiency. These two deaths could have been possibly avoided with the better post-operative support of neonatal intensive care unit. Even so, the survival rate of 54% in our study in the present set of available facilities is comparable with the other local studies.<sup>9</sup>

In summary, bochdalek hernia is one of the main

causes of respiratory distress and cyanosis in newborns and needs early evaluation. The babies who develop symptoms soon after birth (within first few hours) have severe pulmonary hypoplasia and reactive pulmonary hypertension, thus show a poor prognosis. The association of cardiac and multiple malformations affect directly to the outcome of these children. The neonates with late onset of symptoms and less severe associated anomalies have better survival rate. Early diagnosis, better pre-operative and post-operative support and facilities of neonatal intensive care unit will significantly contribute in the reduction of mortality and morbidity among these children. A paediatric surgically oriented educational program for the rural health care providers will help in picking up more cases for the data collection, analysis and better plans for the management in the local surroundings.

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